AMENDMENTS TO THE CLAIMS

This listing will replace all prior versions and listings of claims in the application.

1. (Canceled)

2. (Amended) A **dry powder** pharmaceutical composition according to claim **[[1]] 10.** in which the aromatic flavoring agent is selected from fruit flavoring agents.

3. (Amended) A **dry powder** pharmaceutical composition according to claim **[[1]] 10**, in which the aromatic flavoring agent is a strawberry flavoring agent.

4. (Canceled)

5. (Canceled)

6. (Amended) A **dry powder** pharmaceutical composition according to claim **[[1]] 10**, in which the sodium 4-phenylbutyrate is present in the form of granules which further comprise a binding agent.

7. (Amended) A **dry powder** pharmaceutical composition according to claim 6, which comprises, per 100 parts by dry weight of the composition;

from about 80 to about 90 parts by weight of sodium 4-phenylbutyrate;

from about 3.5 to about 5.0 parts by weight of aspartame;

from about 1.5 to about 3.5 parts by weight of potassium acesulfame;

from about 2.5 to about 5.0 parts by weight of an aromatic fruit flavoring agent; and

from about 3.5 to about 6.5 parts by weight of a binding agent.

8. (Original) A pharmaceutical composition according to claim 7, in which the binding agent is polyvinylpyrrolidone.

9. (Original) A pharmaceutical composition according to claim 7, in which the fruit flavoring agent is a strawberry flavoring agent.

10. (Previously Presented) A dry powder pharmaceutical composition comprising sodium 4-phenylbutyrate, at least one water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and at least one water soluble flavoring agent, the amounts being selected so as to mask substantially the bitter taste and pungent odor of sodium 4-phenylbutyrate.

11. (Previously Presented) A pharmaceutical composition which comprises granules comprising sodium 4-phenylbutyrate, a binding agent, at least one synthetic water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and at least one water soluble flavoring agent, the amounts being selected so that, upon dissolution in water to yield an aqueous solution that contains from about 10 to about 50 mg/ml of sodium 4-phenylbutyrate, the resulting aqueous solution is palatable.

12. (Original) A pharmaceutical composition according to claim 11, which comprises per 100 parts by weight of the composition;

from about 80 to about 90 parts by weight of sodium 4-phenylbutyrate;
from about 3.5 to about 5.0 parts by weight of aspartame;
from about 1.5 to about 3.5 parts by weight of potassium acesulfame;
from about 2.5 to about 5.0 parts by weight of a strawberry flavoring agent; and
from about 3.5 to about 6.5 parts by weight of polyvinylpyrrolidone.

13. (Previously Presented) A concentrated aqueous solution containing at least about 200 mg/ml of sodium 4-phenylbutyrate up about 250 mg/ml, and

having dissolved therein at least one water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and at least one water soluble flavoring agent, the amounts being selected so as to mask substantially, following dilution by at least about 5 fold up to about 10 fold or more with water, the bitter taste and pungent odor of sodium 4-phenylbutyrate.

14. (Original) A concentrated aqueous solution according to claim 13, in which the flavoring agent is selected from fruit flavoring agents.

15. (Original) A concentrated aqueous solution according to claim 13, in which the flavoring agent is a strawberry flavoring agent.

16. (Canceled)

17. (Canceled)

18. (Original) A concentrated aqueous solution according to claim 13, which comprises, per 100 parts by weight of the dry components of the composition;

from about 80 to about 90 parts by weight of sodium 4-phenylbutyrate;
from about 3.5 to about 5.0 parts by weight of aspartame;
from about 1.5 to about 3.5 parts by weight of potassium acesulfame;
from about 2.5 to about 5.0 parts by weight of a fruit flavoring agent; and
from about 3.5 to about 6.5 parts by weight of polyvinylpyrrolidone.

19. (Previously Presented) A concentrated aqueous solution according to claim 14, in which the fruit flavoring agent is a strawberry flavoring agent.

20. (Currently amended) A unit dose for administration to a patient requiring treatment for a urea cycle deficiency according to a regime in which the patient is administered a predetermined number of doses daily corresponding to from about 450 to about 600 mg/kg/day of sodium 4-phenylbutyrate, the unit dose ~~**prepared by diluting with water an aliquot of a concentrated aqueous solution**~~ **containing** **from about 10 to about 50 mg/ml of sodium 4-phenylbutyrate,** ~~**at least about 200 mg/ml of sodium 4-phenylbutyrate up to about 250 mg/ml**~~, at least one water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and at least one water soluble flavoring agent, ~~**the unit dose containing from about 10 to about 50 mg/ml of sodium 4-phenylbutyrate and**~~ the amounts being selected so as to mask substantially the bitter taste and pungent odor of sodium 4-phenylbutyrate.

21. (Original) A unit dose according to claim 20, in which the amount of sodium 4-phenylbutyrate corresponds to no more than about one third of the maximum daily requirement of about 600 mg/kg/day.

22. (Currently amended) A pharmaceutically acceptable aqueous solution ready for administration to a patient requiring treatment for a urea cycle deficiency according to a regime in which the patient is administered a predetermined number of **unit** doses daily corresponding to from about 450 to about 600 mg/kg/day of sodium 4-phenylbutyrate, the solution containing a unit dose of sodium 4-phenylbutyrate, an amount of at least one water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and an amount of at least one water soluble flavoring agent, ~~**the concentration of sodium 4-phenylbutyrate in the aqueous solution ranging from about 10 to about 50 mg/ml and**~~ the amounts of the at least one water soluble sweetening agent and of the at least one water soluble flavoring agent being selected so as to mask substantially the bitter taste and pungent odor of sodium 4-phenylbutyrate.

23. (Previously Presented) A pharmaceutical composition comprising granules comprising sodium 4-phenylbutyrate and a binding agent, the composition further including an effective amount of at least one synthetic water soluble sweetening agent comprising a mixture of aspartame and potassium acesulfame, and at least one water soluble flavoring agent, the amounts of the at least one synthetic water soluble sweetening agent and the at least one water soluble flavoring agent being such that, upon dissolution in water yield an aqueous solution containing from about 10 to about 50 mg/ml of sodium 4-phenylbutyrate to render the resulting aqueous solution palatable to a child.

24. (Original) A pharmaceutical composition according to claim 23, in which the binding agent comprises polyvinylpyyrolidone.

25. (Original) A pharmaceutical composition according to claim 23, in which the flavoring agent is selected from fruit flavoring agents.

26. (Original) A pharmaceutical composition according to claim 25, in which the flavoring agent is a strawberry flavoring agent.

27. (Canceled)

28. (Canceled)

29. (Original) A pharmaceutical composition according to claim 23, which comprises, per 100 parts by dry weight of the composition;

from about 82.5 to about 99.5 parts by weight of sodium 4-phenylbutyrate;
from about 3.25 to about 4.5 parts by weight of aspartame;
from about 1.75 to about 3.25 parts by weight of potassium acesulfame;
from about 3.25 to about 4.5 parts by weight of a water soluble fruit flavoring agent; and

from about 3.25 to about 5.25 parts by weight of polyvinylpyrrolidone.

30. (Original) A pharmaceutical composition according to claim 29, in which the fruit flavoring agent is a strawberry flavoring agent.

31. (Previously presented) A pharmaceutical composition according to claim 23, wherein the granules comprise sodium 4-phenylbutyrate and the binding agent and wherein the granules are mixed with the at least one synthetic water soluble sweetening agent and with the at least one water soluble flavoring agent to form the wetted mass.

32. (Original) A pharmaceutical composition according to claim 23, wherein the granules comprise sodium 4-phenylbutyrate, the binding agent, the at least one synthetic water soluble sweetening agent and the at least one water soluble flavoring agent.

33-63. (Canceled).

64. (New) A method of treating a patient suffering from a condition selected from a urea cycle deficiency and sickle-cell anaemia which comprises administering to the patient in one or more unit doses daily the pharmaceutically acceptable aqueous solution of claim 22.

65. (New) A method according to claim 64, in which the fruit flavoring agent is a strawberry flavoring agent.

66. (New) A method according to claim 64, in which the amounts of aspartame and potassium acesulfame are selected so as not to exceed their respective Acceptable Daily Intakes.

67. (New) A method according to claim 64, in which the solution comprises, per 100 parts by dry weight of the solution: from about 80 to about 90 parts by weight

of sodium 4 -phenylbutyrate; from about 3.5 to about 5.0 parts by weight of aspartame; from about 1.5 to about 3.5 parts by weight of potassium acesulfame; from about 2.5 to about 5.0 parts by weight of a fruit flavoring agent; and from about 3.5 to about 6.5 parts by weight of a binding agent.

68. (New) A method according to claim 67, in which the fruit flavoring agent is a strawberry flavoring agent.

69. (New) A method according to claim 64, in which the unit dose is prepared by diluting with water a concentrated aqueous solution containing at least about 200 mg/ml of sodium 4-phenylbutyrate up the solubility limit thereof measured at 10°C, an amount of at the least one water soluble sweetening agent, and an amount of the at least one water soluble flavoring agent, the amounts being selected so as to mask substantially, following dilution by from about 5 times to about 10 times or more with water, the bitter taste and pungent odor of sodium 4-phenylbutyrate.

70. (New) A method according to claim 69, in which the at least one water soluble flavoring agent is selected from fruit flavoring agents.

71. (New) A method according to claim 70, in which the at least one water soluble flavoring agent comprises a strawberry flavoring agent.

72. (New) A method according to claim 69, in which the concentrated aqueous solution comprises per 100 parts by dry weight of the components; from about 82.5 to about 88.5 parts by weight of sodium 4-phenylbutyrate; from about 3.25 to about 4.5 parts by weight of aspartame; from about 1.75 to about 3.25 parts by weight of potassium acesulfame; from about 3.25 to about 4.5 parts by weight of an aromatic fruit flavoring agent; and from about 3.25 to 5.25 parts by weight of a binding agent.

73. (New) A method according to claim 72, in which the binding agent comprises polyvinylpyrrolidone.

74. (New) A method according to claim 64, in which the patient is administered three unit doses daily and in which the amount of sodium 4-phenylbutyrate in the unit dose corresponds to no more than about one third of the maximum daily requirement of about 600 mg/kg/day.

75. (New) A method according to claim 72, in which the condition is a urea cycle deficiency.

76. (New) A method according to claim 72, in which the condition is sickle-cell anemia.